



# Dr. Kimberley Tan

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Sydney Children's Hospital, Randwick





## Initial presentation

Well 5 year old boy

No Past Ocular history

School screening detected poor vision right eye

Intelligent boy, excellent motor skills, normal development

- Rare mild headache – “likes it quiet”
- No headache on waking, no vomiting
- Sleeps well, no snoring
- Head size – growing normally: 51.5cm (50<sup>th</sup> centile)
- No recent medication



## Initial Ophthalmic exam

VAR 6/36 VAL 6/6

Near: R N12 L N5

Healthy, **not** overweight, exceptionally co-operative boy

Refraction R: +3.5+0.25x85 L: +0.5

No strabismus, full ocular rotations

Ishihara plates (held very close RE) - normal

Media clear

Confrontation fields: full



## Initial Ophthalmic exam – discs





# Investigations

**MRI Brain - Normal**

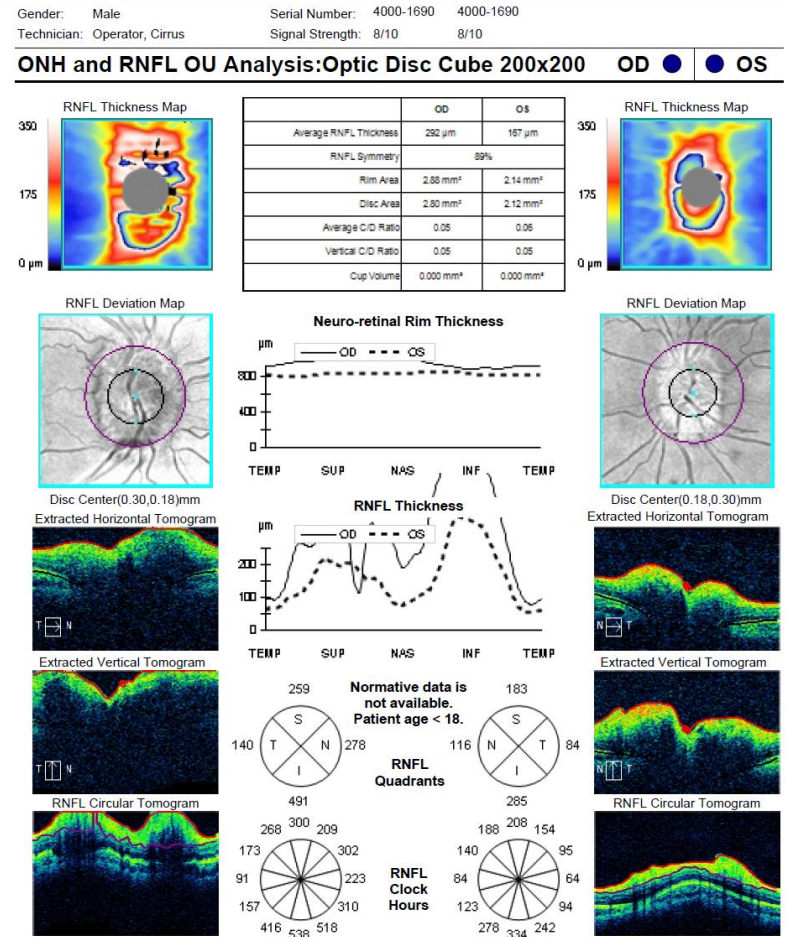
**B Scan Optic nerves - Normal, no drusen**

**HVF - Within Normal Limits for age**

**OCT - Not c/w drusen**

**Significant Nerve Fibre Layer (NFL) swelling**

**Autofluorescence - Normal**





## Anisohypermetropic amblyopia

“Swollen Discs” – incidental finding

- Congenital disc elevation (Hyperopia)

Vs

- True disc pathology: Papilloedema or papillitis



# Treatment

Glasses

Amblyopia therapy – patching LE

Review

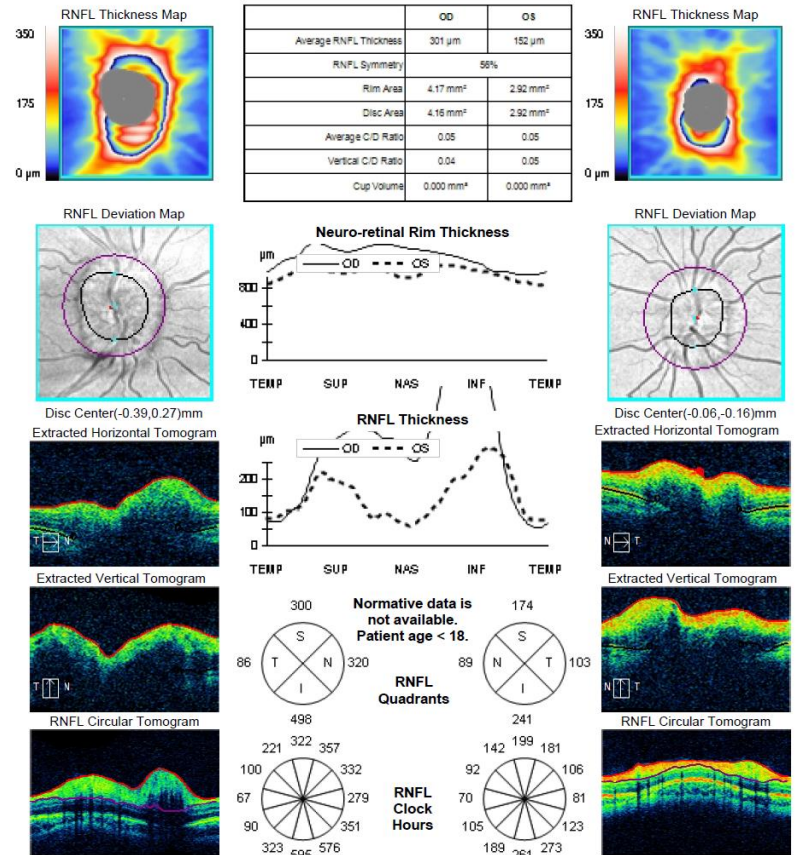
Vision improved 6/19 RE , 6/6 LE

Discs unchanged

Repeat OCT

Gender: Male      Serial Number: 4000-1690    4000-1690  
 Technician: Operator, Cirrus      Signal Strength: 7/10    10/10

## ONH and RNFL OU Analysis: Optic Disc Cube 200x200    OD ●    ● OS





## Further investigation

### Involved Paediatric Neurologist

- Complete Neurologic examination otherwise normal
- Routine Bloods: normal, not anaemic, Iron studies normal
- Lumbar Puncture
  - Nitrous oxide sedation
  - Opening Pressure 13.5cm H<sub>2</sub>O
  - No cells, normal biochemistry





# Progress

Continue glasses & patching

Vision slowly improving

Progress OCT monthly – no major change over months

No change to optic nerve appearance



## 1 year later (now 6 years old)

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VAR 6/12 VAL 6/6

Refraction unchanged

Stereo 100 secs

Systemically well

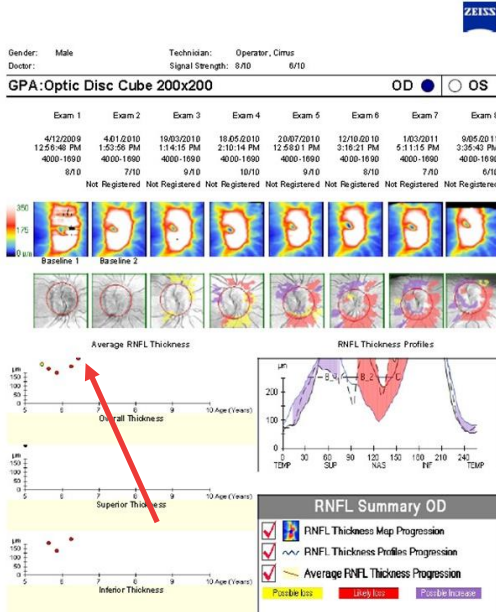
HVF – normal

Discs appear more swollen

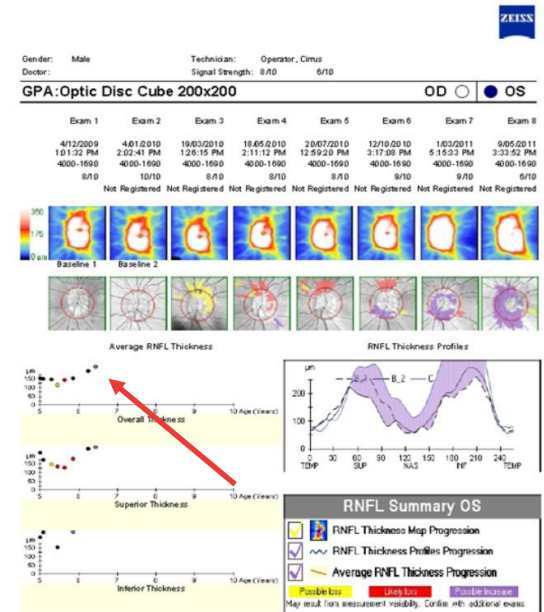
OCT definite, repeatable NFL increase in swelling

# 1 year later (now 6 years old)

## Right Eye



## Left Eye





# Repeat assessment as disc swelling worse

**Repeat MRI / MRV - normal**

**Repeat neurologic exam – normal**

**Repeat Lumbar Puncture**

- Nitrous Oxide
- Opening Pressure 18 cm H<sub>2</sub>O
- Normal cells / biochemistry

PLEASE ANSWER THE  
QUESTIONS POSED ON THE  
NEXT SLIDE BEFORE  
PROCEEDING WITH THE CASE  
PRESENTATION



# HOW WOULD YOU DEAL WITH SUCH A PATIENT?

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1. What investigation would you choose next:
  - a) CSF infusion study
  - b) Formal ICP monitor
  - c) Sleep study
  - d) Visual Electrophysiology
  - e) Dural Venous Sinus Manometry
  - f) None for now
  
2. Would you recommend:
  - a) Oral Diamox
  - b) Oral Topamax
  - c) Corticosteroids
  - d) Optic nerve sheath fenestration
  - e) Neurosurgical shunt
  - f) CPAP
  - g) Dural Venous sinus stenting
  - h) No treatment – continue to observe

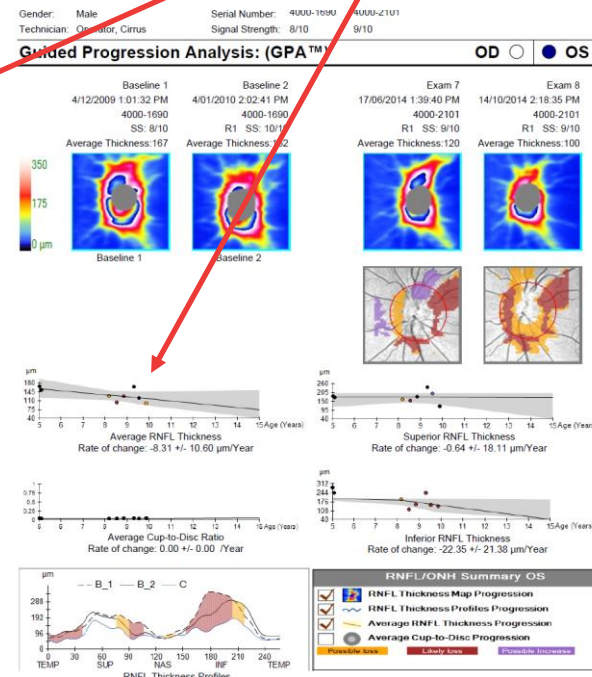
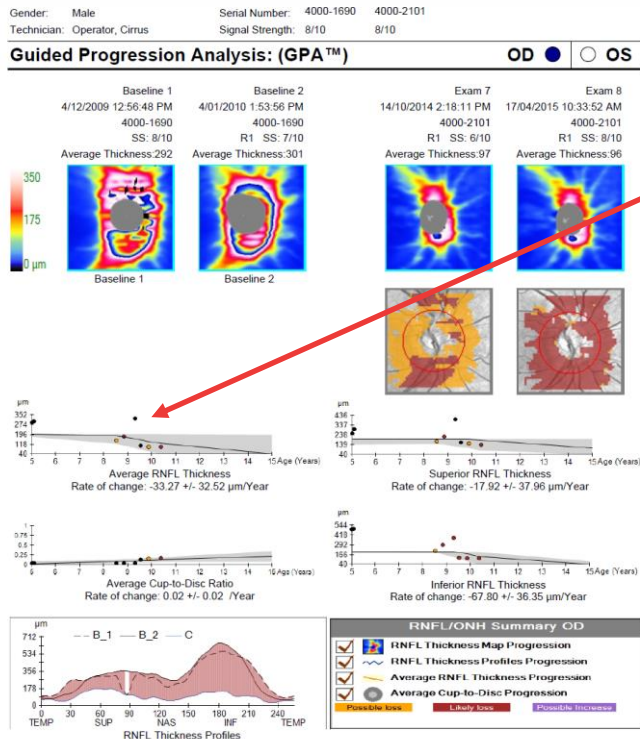


# What Next?

## Diamox trial

- immediate NFL improvement clinically and on OCT
- variable course but slow wean over 4 years

Treatment commenced



# The patient is now 11 years

Total 5 year course – now off Diamox

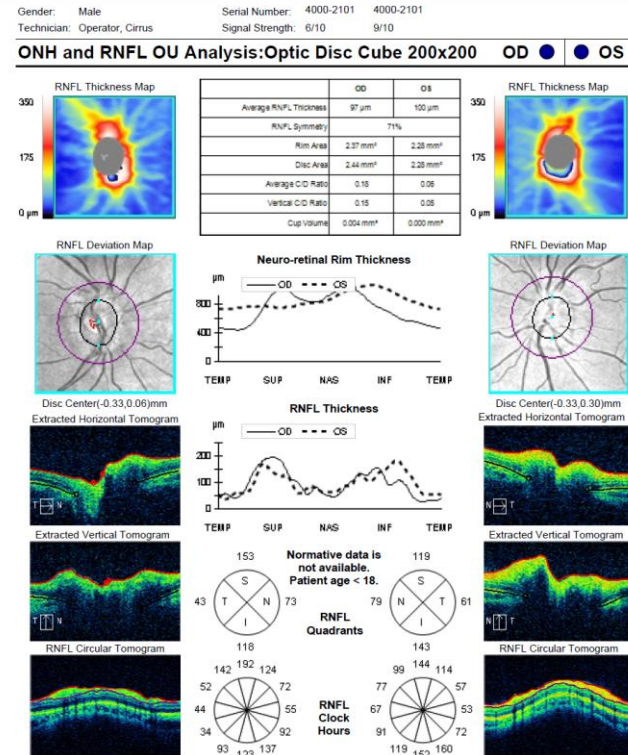
Well, normal development, no headaches

No optic neuropathy

Normal fields/colours

Mild RE anisometropic amblyopia

VAR 6/9 VAL 6/6







## COULD OUR EXPERTS PLEASE ANSWER THESE QUESTIONS IN ADDITION TO THOSE ASKED EARLIER?

1. What do you feel is the aetiology to his optic nerve swelling and what other investigations would you have liked in his workup?
2. Please explain the value you place on a standard Lumbar Puncture in these situations? And do you ask for any special instructions in the way it is performed? (Anaesthetics / nitrous / position etc). Do you prefer CSF Infusion studies and / or formal ICP monitoring?
3. How long do you follow these children for? When is it safe to discharge from regular follow up?



*Thank you!*

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